

Evaluation of Total Repair of Complete Atrioventricular Septal Defect In infants and Children

Thesis

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LIST OF ABBREVIATIONS

ABG	Arterial blood gases
ACT	Activated clotting time
ASD	Atrial septal defect
AVSD	Atrio-ventricle septal defect
AVV	Atrio ventricular valve
CAVC	Complete atrio-ventricular canal
CHD	Congenital heart disease
CXR	Chest x-ray
DS	Down syndrome
ECG	Electrocardiogram
ICU	Intensive care unit
IQR	Interquartile range
LAVVR	Left atrio-ventricular valve regurgitation
LIL	Left inferior leaflet
LSL	Left superior leaflet
LVOTO	Left ventricle outflow tract obstruction
MR	Mitral regurgitation
PA	Pulmonary artery
PPHN	Persistent pulmonary hypertension of newborn
PVOD	Pulmonary vascular obstructive disease
SD	Standard deviation

LIST OF ABBREVIATIONS (CONT.)

VSD	Ventricular septal defect
PDA	Patent ductus arterioses
CPB	Cardio-pulmonary bypass
LV	Left ventricle
RV	Right ventricle
ECMO	Extra corporal membrane oxygenator
PAP	Pulmonary artery pressure
PAB	Pulmonary artery banding
Sig.	Significant
Ns.	Non significant

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Introduction

CAVC is a complex cardiac malformation characterized by a variable deficiency of the atrioventricular area (crux cordis) in the developing heart. The malformation involves the atrial, ventricular and atrioventricular septa and both atrioventricular valves (*Calabrò and Limongelli, 2006*).

The lesion is a deficiency of the tissues that, in the normally structured heart, interpose between the atrial and ventricular chambers. In the normal heart, there is but one atrioventricular septum. This partition is a component of the normal aortic root and is interposed between the cavities of the left ventricle and the right atrium. It was believed initially that a muscular atrioventricular septum was also present in the normally structured heart (*Anderson et al., 2010*).

CAVC accounts for about 3% of all cardiac malformations. Atrioventricular canal occurs in two out of every 10,000 live births. Both sexes are equally affected, with a slightly higher frequency in female (female/male ratio 1.3/1) and a striking association with Down syndrome was found about 70% are children with Down syndrome. Of children with Down syndrome, about 40% have congenital heart defects and 50% of the defects are AVSD (*Kim et al., 2001*).

Half of children with untreated CAVC die in the first year of life. The main cause of death in infancy is either heart

failure or pneumonia. In surviving patients with unrepaired complete atrioventricular canal, irreversible pulmonary vascular disease becomes increasingly common, and affects virtually all patients older than 2 years of age. Long-term prognosis in patients with irreversible pulmonary hypertension is poor (*Trachte et al., 2005*).

The complete form of CAVC shows an ostium primum atrial septal defect, a common atrioventricular valve and a variable deficiency of the interventricular septum inlet. (*Boening et al., 2002*).

This anatomic arrangement gives a scooped out appearance to the ventricular inlet and a long and narrow morphology to the left ventricular outlet. The key finding for the anatomic classification in type A, B or C of this malformation is the morphology of the common atrioventricular valve (*Digilio et al., 1999*).

The first repair of complete atrioventricular canal (CAVC) by *Dr. Walton Lillehei* in *1955* performed using controlled cross-circulation, involved suture closure of the atrial septal defect (ASD) and ventricular septal defect (VSD) with direct suturing of the atrioventricular valve (AVV) leaflets to the crest of the interventricular septum. This practice was subsequently abandoned in favour of placement of a patch for closure of the ASD and VSD components (*Najm et al., 1997*).

Many surgeons during the 1970s and early 1980s advocated palliation with pulmonary artery banding to delay surgical repair beyond infancy.

On the other hand, as operative techniques and postoperative care improved as well as cardiopulmonary bypass in infants became safer, most centres abandoned this practice and opted for elective repair before 6 months of age in asymptomatic infants with complete AV canal defects (*Stellin et al., 2003*).

Two standard methods for repair of CAVC defect are in use today. In the traditional single-patch technique, the same patch is used for closure of the ASD and VSD; the AVV leaflets are re-suspended onto this single patch, with a need for leaflet division in order to allow patch placement for Rastelli type C, which by definition has a common superior leaflet. In the double-patch technique, 2 separate patches are used, one for closure of the VSD and the other for closure of the ASD; the AVV leaflets are typically not divided (*Jonas and Bassem 2010*).

A primary characteristic of the modified single-patch technique, which was introduced in the mid-1990s, is the obliteration of interventricular communication by direct closure of the bridging leaflet and septal crest (*Jeong et al., 2009*).

Aim of the Work

To evaluate the early results of surgical total repair of complete atrioventricular septal defect. In relation to three important risk factors:

- a) Early repair in the first year of life versus later repair.
 - b) Mild to moderate pulmonary hypertension (less than half the systemic Pressure) versus severe pulmonary hypertension.
 - c) Primary repair versus repair after previous pulmonary artery banding.
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